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Oral Rehabilitation of Patients with Hypohidrotic Ectodermal Dysplasia: When, Why and How to Intervene

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Authors' contributions

This work was carried out in collaboration between all authors. Author II designed the study, produced the outline, wrote the parts of the text dealing with the orthodontic considerations and provided the photographs. Author EK wrote the sections of the text related to the prosthodontic management, proofread and corrected the manuscript. Author KM performed the literature search and cooperated in the writing. All authors read and approved the final manuscript.

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ABSTRACT

Hypohidrotic ectodermal dysplasia (HED) patients present absence of many permanent teeth, alveolar deficiency and severely affected maxillofacial skeleton. The management commonly includes a series of removable dentures and orthodontic appliances, adapted constantly to the child's growth and development. The dental literature lacks a protocol to elucidate when, why and how to intervene for the modification of jaw growth pattern and for the treatment of dental and occlusal problems to maximize the clinical therapeutic outcome. The aim of the present review was to propose and redefine the treatment plan for the orofacial rehabilitation of patients with HED.
Normal craniofacial growth is described, followed by a review of published findings about facial

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growth in HED patients. Subsequently, the treatment strategy is presented, focusing on the stages of physical growth and dental development: (a) infancy and early childhood (primary dentition years), (b) late childhood (mixed dentition years), (c) adolescence (early permanent dentition years) and (d) adulthood (permanent dentition years), and to the special characteristics related to each developmental stage. Finally, the treatment possibilities and restrictions are considered, and a protocol defining when, why and how to intervene is proposed.

Keywords: Hypohidrotic ectodermal dysplasia; rehabilitation; oligodontia; multidisciplinary; prosthodontics; orthodontics.

1. INTRODUCTION

Hypohidrotic ectodermal dysplasia (HED) is a congenital syndrome inherited as an X-linked recessive trait [1,2]. It is characterized by abnormalities of tissues of ectodermal origin namely skin, nails, hair and teeth. Individuals with HED exhibit specific facial features, such as frontal bossing, concave depressed middle face, depressed nasal bridge, everted lips, prominent chin, reduced facial height and facial depth. In addition, there is an absence of many deciduous and permanent teeth manifested as oligodontia (congenital lack of more than six permanent teeth) or even anodontia (congenital lack of all permanent teeth). Teeth usually present are the maxillary incisors, maxillary and mandibular first molars and maxillary canines, which often have abnormal crown shape, short roots and large pulpal chamber [3-10]. The severely affected maxillofacial skeleton contributes to the patients' typical 'aged-face', even during growth. Additionally, the absence of a large member of teeth leads to deficient alveolar growth at edentulous sites, impaired masticatory function, and severe disturbances of the stomatognathic system. Chewing and swallowing dysfunction, dryness of the mouth and speech difficulties are frequent [11].

The facial and oral features, affecting esthetics and function, may have a negative psychological effect in the patients with HED. The child's self-image is usually complete by age 4-5 years [12] and the severely affected child is prone to psychosocial and psychological problems [13,14].

Treatment for children with HED is multidisciplinary. Prosthodontists and orthodontists are particularly concerned. The dental management is usually realized by a series of removable partial dentures (RPDs), complete dentures (CDs) and removable or fixed orthodontic appliances during the growth period until the definitive rehabilitation at the end of growth [8,11,15-17]. It is an active process, constantly adapted to the child's growth and development, and still remains a challenge for every clinician involved in the treatment of patients with HED. When, why and how to intervene and what protocol must be followed to modify the growth pattern and treat the dental and occlusal problems, in order to maximize the overall treatment outcome? In response to these critical questions, the aim of the present review was to discuss and redefine the treatment plan for the orofacial rehabilitation of patients with HED, with emphasis on prosthodontic and orthodontic aspects, which have emerged during the authors' long-term collaborative work on the issue.

2. GROWTH OF CRANIOFACIAL COMPLEX IN HED PATIENTS VERSUS NORMAL GROWTH

Knowledge of craniofacial growth features of HED individuals is essential, in order to include them among the diagnostic criteria for treatment planning. Thus, it is primarily important to be familiar with the normal growth of different parts of craniofacial complex and the directional changes occurring in each part.

2.1 Normal Facial Growth- General Considerations

2.1.1 The cranial base

Growth of the cranial base is the result of endochondral growth through synchondrosis within and between the ethmoid, occipital and sphenoid bones. The intraoccipital synchondrosis closes before 5 years of age, whereas the intraethmoidal and the intrasphenoidal close before birth. The sphenoethmoidal synchondrosis closes at around 6 years and that segment, designated as anterior cranial base, becomes relatively stable early in life. Therefore, the anterior cranial base is used for superimpositions on lateral cephalometric radiographies to evaluate changes in the face that occur during growth or treatment. The sphenooccipital synchondrosis closes at around 13-15 years of age and any changes in the flexure of the cranial base result from surface bone deposition and resorption. Extreme changes in the cranial base flexure significantly influence other parts of the craniofacial complex that are related to the cranial base, either directly or indirectly [18].

2.1.2 The maxillary growth

Maxillary growth in width occurs mostly at the midpalatal suture during the first 5 years of life. Thereafter, any additional increase in the width of the anterior maxilla occurs as a result of bone deposition on the outer surfaces of the maxilla and of the buccal eruption of the permanent teeth. During growth, the maxilla is translated downward and forward, at an extent that depends on both the cartilage of the nasal septum and the surrounding soft tissues. During growth, individuals may have varying maxillary growth potentials related to anterior cranial base, and may present extreme variations such as pure horizontal growth or pure vertical growth [18,19].

2.1.3 The mandibular growth

Mandibular growth is expressed as a downward and forward displacement, while the growth at the head of the condyle occurs in an upward and backward direction. The growth at the condyles compensates for the vertical displacement of the mandible and accommodates for the eruption of the teeth vertically. Bone resorption at the anterior border of the rami accounts for the anteroposterior growth of the rami and body of the mandible. These changes increase the posterior length of the mandible to accommodate the erupting permanent molars. Condylar growth can vary between a sagittal (backward and upward) and vertical (forward and upward) direction and this variation influences the mandibular growth direction [18-20].

2.1.4 The growth of the alveolar processes

Growth of the alveolar processes is solely dependent on the presence and eruption of teeth. The bone of the alveolar process exists only to support the teeth. If a tooth fails to erupt, alveolar bone never forms in the area it would have occupied. The growth of the maxillary and mandibular alveolar processes, closely linked with the teeth eruption, increases the vertical height of the face [18,19].

Growth changes of the various parts of the craniofacial complex occur simultaneously and are interdependent. As there is a significant amount of individual variations in amount, direction and rate of growth, it is difficult for the clinician to predict it. The overall direction of growth for each part of the craniofacial structures, as well as the pattern of growth can be evaluated by the analysis of lateral cephalometric radiography [18,19].

2.2 Craniofacial Growth Changes and Physical Growth and Development

In infancy, there is normally a rapid pace of body growth, which declines by early childhood and stabilizes at a moderate level during late childhood. During adolescence, there is a growth spurt with major changes in body development and then, at adulthood, the growth ceases [19].

The growth of the jaws correlates with the physiologic events of puberty in about the same way as growth in height. During normal growth there is an internal and an external rotation of jaws which result in variations in jaw orientation and face development. An excessive forward rotation of the mandible results in a low anterior lower face height, whereas an excessive backward rotation of the mandible results in a high anterior lower face height, producing short face and long face individuals correspondingly [19]. During the adolescent growth spurt, there is a significant growth in length of mandible and a growth at the sutures at the maxilla, milder than in mandible. The acceleration in mandibular growth in relation to the maxilla produces differential jaw growth, thus the face becomes less convex, as the mandible and the chin become prominent [19].

Growth in length and height of the jaws continues through puberty. Growth in width of the jaws, including the width of the dental arches, tends to be complete before the adolescence growth spurt. Vertical growth is completed first, followed by growth in length and, finally, in height. In both sexes growth in height of the face lasts longer than growth in length [19]. However, many physical changes differ between girls and boys. Puberty and adolescence growth spurt occur on average nearly 2 years earlier in girls than in boys. Thus, implementation of treatment must be done earlier in girls than in boys to take advantage of the adolescence growth spurt [19].

2.3 Facial Growth in HED Patients

Literature concerning the facial growth pattern in HED patients invoked the observed differences in comparison with healthy control groups. The cephalometric analyses revealed a unique abnormal craniofacial development, a reduced anterior face height and significantly reduced to 24.07±0.97º (normal: 26±4º) inclination of the mandible [21]. An abnormally short maxillary depth and strongly reduced lower facial height are also reported [22]. Anthropometric and cephalometric measurements demonstrate reduced maxillary length, normal size and shape of the mandible and reduced sagittal jaw relationships [23].

Longitudinal studies concerning the pattern of facial growth in HED patients are limited, but highlight the vertical and sagittal deficiencies during craniofacial growth and development. Bondarets et al. [24], analyzing lateral cephalometric radiographs of HED patients found a tendency for the individuals to develop a marked Class III sagittal jaw relationships with time. In addition, they demonstrated a significant difference in growth between the anterior and posterior face heights, suggesting that their subjects had a tendency for anterior growth rotation.

Studies on facial tissues also describe differences between normal reference subjects and HED patients [21,23,25,26]. Soft tissue alterations affect patients with HED during growth as a consequence of the reduced vertical height. Cephalometric studies reported that the subnasion point was behind the aesthetic line (EL), the naso-labial angle became acute and lips were protuberant and everted [21]. Short nose, reduced facial convexity and protruding lips in men and retruded lips in female carriers characterize also the soft tissue profile [23]. One cephalometric study which compared the HED sample to a matched non-syndromic control sample revealed a reduction in facial soft tissue thickness in HED children [22]. Dellavia et al. [9] collected 3-D coordinates of facial landmarks to estimate the volumes of forehead, nose, upper and lower lips, maxilla and mandible separately for the childhood growth period and the adolescence. They found that the nose had reduced growth at all times, the global growth of the lower lip was greater in HED subjects, the global facial growth was slightly reduced compared with normal peers and the peak of development of both jaws was delayed by approximately two years towards later adolescence.

Facial growth deficiencies in HED patients have been examined in relation to the degree of hypodontia. Findings of a longitudinal study in HED patients who had not undergone any orthodontic or prosthodontic treatment [24] suggest that the retrognathic maxillary tendency and the vertical deficiency are most likely due to the dental and functional compensation and not to altered growth pattern. In another longitudinal study [9], in which the patients had been using prosthodontic and orthodontic appliances, the authors found a trend of growth nearly double than that of the non-rehabilited HED patients, confirming the normal growth potential of HED patients and underlining the ability of prosthodontic and orthodontic appliances to modify maxillary and mandibular growth.

Therefore, it seems that the deficiencies existing in sagittal and vertical dimension in HED patients could be explained by the high number of missing teeth. As mentioned, normally the increase in the vertical height of the face is the result of the growth of the maxillary and mandibular alveolar processes which is closely linked with the eruption of teeth. In HED patients, the extensive lack of teeth affects the growth of the alveolar processes, limiting the alveolar bone growth, which in turn results in impaired sagittal and vertical growth. The more severe the dental agenesis, the more evident the maxillary retrusion, the mandibular protrusion and the vertical deficiency [27].

3. ORAL REHABILITATION OF HED PATIENTS: GENERAL CONSIDERA TIONS AND CLINICAL PROCEDURES

Oral rehabilitation of HED patients requires a multidisciplinary approach to treatment planning and execution. The therapeutic strategy depends on age, severity of dental agenesis and degree of malformation. It varies by case and needs parents' consent and patients' collaboration. There are different therapeutic choices, such as removable partial prostheses, complete dentures, fixed partial dentures, orthodontics, implants and combinations of them.

In order to establish a diagnosis and provide a treatment plan, the diagnostic tools usually used are diagnostic casts, as well as panoramic and lateral cephalometric radiographs. Panoramic radiographs reveal the congenitally missing primary and permanent teeth in maxillary and mandibular arch, their root form and stage of development. Lateral cephalometric radiographs are used to evaluate the direction of craniofacial growth (horizontal, average or vertical), the position and size of the jaw bases in relation to anterior cranial base, the sagittal and vertical base relationships, the morphologic particularities of mandible and its growth pattern, and the position and the axial inclination of the incisors [28].

4. TREATMENT PROTOCOL - WHEN, WHY AND HOW TO INTERVENE IN HED PATIENTS

Generally, the treatment of HED patients should be instituted early, should continue during the whole growth period and should finish with patients' skeletal maturity. It is a long and continuous struggle against the defects that the patients present in craniofacial growth and development. Moreover, it has to be a continuous and active process, aiming to enhance or develop normal speech, to maintain or modify the growth pattern, to support jaw relationships and temporomandibular joint function, and to improve the esthetics from the youngest ages until adulthood [4,8].

Typically, treatment begins with the fabrication of a removable partial or complete prosthesis at an early age. During the mixed dentition stage, the prosthesis is modified and replaced when needed to accommodate growth changes and erupting teeth, and in permanent dentition stage it is replaced by a definite restoration, preferably fixed, depending on the number and position of the permanent teeth and implant placement potency [15,16,29-32]. It is presently unclear at what stage of physical and facial growth and development the treatment must be more or less active, at what stage prosthodontics and/or orthodontics are more efficient to modify facial growth or contribute better to the patients' needs. Each growth period has its proper characteristics which have to be considered in order to define the appropriate appliance in the appropriate time to increase the efficiency of the appliances and the treatment outcome. Based on the sequence of growth phases, the authors have in consultation designed a concise protocol of the proposed interventions, to serve as a guide for the combined treatment plan (Table 1).

During the three main growth periods before maturity, there are opportunities and limitations which should be carefully considered.

4.1 Infancy and Early Childhood - The Primary Dentition Years

Patients diagnosed with HED are sometimes referred very young, at age 2 or 3, for treatment. At that age parents are not primarily concerned about facial esthetics as there are yet few signs of facial imbalance. Their main priority is to assure that their children are able to eat and function like their peers. Most often the patients are referred at age 4-5 years. At this time children exhibit rapid rates of growth and the anteroposterior and vertical problems in HED patients become more apparent. The mandible often begins to attain a forward position and the jaw discrepancy begins to install. For clinicians, the target at this early age is primarily to assure eating, speech and swallowing. Very young children need to normalize these functions which gradually will contribute to monitoring and preventing the expected growth disturbance [30,32].

According to the number of existing teeth, RPDs or CDs can be constructed to substitute the missing teeth, to improve the masticatory function, the speech and esthetics. At the same time the dentures serve to maintain the vertical facial height and normal anteroposterior jaw relationships preventing the anterior displacement of the mandible. The vertical height can be maintained by constructing dentures according to the patients' occlusion, dictated in part by the occlusal contacts of the existing teeth [29]. However, in many of the children with oligodontia, the vertical dimension of occlusion (VDO) appears reduced, probably because of the lack of enough opposing teeth. This reduction is manifested by the appearance of the lower face, and often contributes in severe reduction of the vertical space available for the denture. A modest increase of the VDO, estimated according the prosthodontic criteria, is often desirable, in order to achieve better aesthetics, speech and support of the perioral and masticatory muscles' functional length. Such an increase can be incorporated in the prosthodontic device, but has to be closely inspected, because removable appliances are not typically recommended to support increase of the VDO [27]. Another problem of HED patients, is the tendency for a forward rotation of the mandible, due to the severe lack of teeth.

Table 1. Combined prosthodontic/orthodontic protocol for treatment of HED patients

Growth period	Dentition stage	Prosthodontics	Orthodontics	Surgery
Infancy and early childhood (Rapid growth rate)	Primary dentition	Partial/complete dentures with 3-direction jackscrew Partial/complete dentures with 2-direction jackscrew Implants in selected cases	Partial/complete dentures with jackscrew	Implant surgery in selected cases
Late childhood (Low growth rate)	Mixed dentition	Partial/complete dentures with 3-direction jackscrew Partial/complete dentures with 2-direction jackscrew	Removable orthodontic appliances with jackscrew Functional orthopedic appliances (face mask, chin-cup)	
Adolescence-time of puberty (Rapid growth rate)	Early permanent dentition	Partial/complete dentures with 3-direction jackscrew	Functional orthopedic appliances (face mask, chin-cup) Bite plates Fixed appliances	
Adulthood (Residual growth- low rate)	Permanent dentition	Partial/complete dentures Implant prosthodontic restorations	Fixed appliances	Bone augmentation Implant surgery Orthognathic surgery combined with orthodontic treatment

To overcome this, the authors advise the construction of dentures with a relatively excessive overbite. RPDs, CDs or orthodontic removable appliances constructed to provide accentuated overbite guide the mandibular position and prevent the anterior shifting. However, such an occlusal scheme has to be implemented with caution, as it may induce unfavorable horizontal loading of the dentures in function, predisposing for loss of stability [33].

No consensus exists on the ideal age for beginning prosthodontic rehabilitation. According to published case reports, removable dentures have been constructed and effectively used for children as young as 3 years [32,34-37]. A 2010 case report describes the process and 3-year follow-up of a set of removable prostheses constructed for a 2-year old boy suffering from ED, and stresses the corrective effect on the prognathic tendency [38]. Substantial esthetic, functional, and psychological benefits can be gained by the provision of complete dentures in very young HED children [39], however, because of the removable nature of the prostheses it is absolutely necessary for the children to be willing to use them. The authors' personal experience, in accordance to the developmental psychology data, is that children are not usually willing to accept their removable teeth substitutes before the age of the first school years, at about 5 or 6 years of age [40]. By this age, the evolving need to be similar with their peers contributes to their willingness to accept the burden of both the construction process and wearing of the denture. Before that, the child is introduced to the prosthodontist, and his/her attitude is assessed by their respond to the typical diagnostic procedures, especially impression making and maxillomandibular relationships recording. Even if the child proves to be unwilling to cooperate, these introductory meetings contribute in the establishment of a trustful relationship between the clinician and the patient, and his/her parents, which is the basis for their cooperation for the years to come [41].

In non-cooperative children implants have been reportedly placed in very young age. Smith et al. [42], and Guckes et al. [43], placed one and six implants respectively in their two male patients, aged 5 and 3 years, along with a removable superstructure and followed them for 5.5 and 5 years respectively. Alcan et al. [44], inserted four implants in a 4-year old boy and constructed a fixed prosthesis which restored only the six mandibular anteriors. After 6 years the prosthesis was still functional, but the occlusal relationship, as well as the patient's vertical growth pattern had been changed. As animal studies have been shown that implants tend to behave as ankylosed teeth and not follow the growth changes [45-47], implant placement in very young children is not generally encouraged.

For cooperative children, removable prostheses inserted as early as possible improve oral function, support the vertical height and prevent abnormal mandibular posture during growth. The vertical and anteroposterior impairment of HED patients can be overcome with removable prostheses, provided that the dentures maintain proper vertical and anteroposterior relationships [30]. To achieve this goal, the dentures have to be comfortable and stable (Fig. 1).

Fig. 1. Intraoral photographs of a 5-year-old boy with HED. (a, b) View of occlusion with reduced VDO and excessive vertical overlap, (c, d) intraoral view of maxilla and mandible, and (e) the occlusion after the placement of a mandibular RPD with wrought wire clasps and an acrylic plate for bilateral occlusal coverage of posterior teeth. (Patient of the Postgraduate Clinic of Orthodontics, treated by the authors)

The authors' long-term engagement in children prosthodontics has shown that stability is not an easy task to achieve for such removable appliances. Stability problems arise from the small dimensions of childrens' oral cavity, from the extremely limited denture foundation area, which stems from the severely limited (virtually non-existing) alveolar crest, and from the lack of saliva, which is a crucial retentive element for removable prostheses. The jaw relations discrepancies attenuate the stability problems, as they result in unfavourable biomechanical conditions, and the exaggerated interarch distance further contributes to such problems. For RPDs, the conical shape and small size of available abutment teeth, as well as the need to keep them intact and to avoid crown modifications in order to preserve them hinders the establishment of a proper support and retention scheme [27]. Modern composite materials with high adhesive qualities have been proved an effective means to build-up the crown of the natural teeth, thus improving, in a non-invasive manner, both aesthetics and denture retention. Dentures are usually entirely tissue borne, retained by wrought wire clasps which provide inadequate retention. Moreover, there is an increased risk of soft tissue irritation. As is well exercised in adult patients' dentures, the denture periphery, external surfaces and teeth position should be designed and shaped in harmony with the surrounding musculature, and use it for stabilizing the prosthesis during function [30]. Finally, modern denture aids, such as denture adhesives [38] and soft liners can be used to enhance retention. The prosthodontist's great ally against the above drawbacks is the children's great adaptation potential.

Well-fitted dentures ensure proper vertical and anteroposterior relationship, but at the same time they may inhibit physiological growth of jaw and fail to support the transversal growth. The authors' clinical experience has shown that all prosthodontic appliances placed at this developmental stage must be reinforced by a 3-directional jackscrew in the maxilla and/or a 2-directional jackscrew in the mandible in order to not only accommodate, but also enhance the growth of alveolar processes in the 3 dimensions of space. With regular opening of the screw (0.25 mm/week), growth in anteroposterior, vertical and transversal direction can be readily monitored, better dentoskeletal conditions are created and a more harmonious environment for the subsequent phases of the treatment can be established. From the prosthodontic aspect, however, the jackscrew addition abates the prosthesis rigidity and further complicates the function, as it permits unilateral independent movement of its parts across the midline [48]. Regular recalls, at about 2-month intervals, are absolutely necessary in order to keep control of the prosthesis function and monitor the growth of the oral tissues (Fig. 2).

4.2 Late Childhood - The Mixed Dentition Years

The goal of the treatment in the late childhood years is to maintain and monitor the oral function. Growth is less rapid than in early childhood, but the progress of vertical, sagittal and transversal facial growth should be monitored; as natural deciduous teeth are gradually lost, esthetic needs appear and need to be addressed.

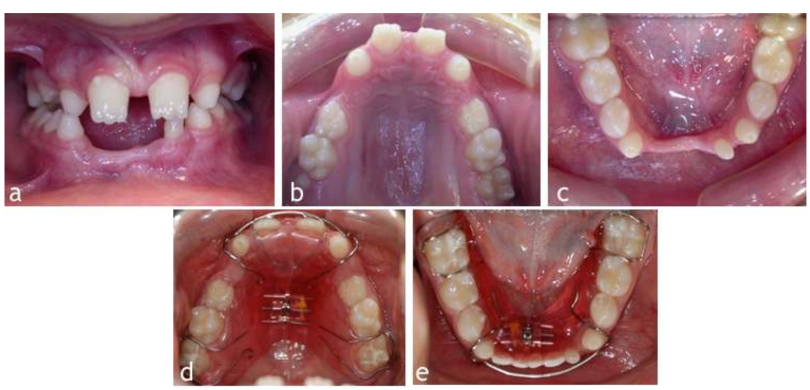

Fig. 2. Intraoral photographs of a 7-year-old boy with HED. (a) View of occlusion showing the restricted transversal growth and the anomalous development of anterior mandibular alveolar ridge, (b, c) view of maxilla and mandible, and (d, e) view after the positioning of orthodontic appliance with a 2-directional jackscrew at the maxilla and the mandible. (Patient of the Postgraduate Clinic of Orthodontics, treated by the authors)

In young HED patients simple, provisional removable dentures are the restoration of choice as they can readily respond to the changes of the oral environment with regular follow-up, adjustments to accommodate exfoliating or erupting teeth and replacements as dictated by the growth of the jaw bones [9,31,49-54]. Replacement dentures at this stage are constructed based on the acquired experience about the specific oral conditions and anticipated problems. Existing prostheses can be used to induce VDO changes or as impression trays. The patients become readily adjusted to the new dentures, as they have already developed the necessary skills and gradually mature into skillful denture users [30]. Once again the authors highlight the necessity to reinforce the prostheses by a 3-directional jackscrew in maxilla and 2-directional jackscrew in the mandible. The opening of the screw at this time of minor physical growth, should be regular but less frequent, at approximately 1 mm/month.

Osseointegrated implants have also been proposed. Kramer et al. [55] report the case of a 8-year old boy with HED who was fitted by 2 implants in the anterior mandible and removable dentures which served satisfactory for 2 years. They therefore recommend implant placement at this age, but also caution about the several issues, such as implant site and patient compliance, that need to be considered. Bergendal et al. [56], McMillan et al. [57], Martin et al. [58] and Aydinbelge et al. [59] have also published case reports of implant placement in a 6- to 8-year-old HED patients, and report satisfactory results for up to 4 years.

Despite the occasional positive results, serious reservations have been expressed concerning the outcome of early implant insertion. Guches et al. [60] conducted a prospective clinical trial to assess the fate of implants placed in ED patients and reported 85% survival for up to 6.5 years. They concluded that implant placement in growing individuals is not a routine procedure and its timing should be decided through a cost-benefit assessment. A similar approach is evident in the case series reported by Lauwers et al. [61], who emphasize the need for a systematic multidisciplinary treatment. After following the behavior of implants placed in growing jaws, a Swedish group [45-47] reported that implants behave as ankylosed teeth. In their series of publications, the group of Cronin and Oesterle [62-66] suggest that the placement of implants in the growing maxilla should be avoided before 15 years of age. In Sweden, implant-supported prostheses had been a rare treatment modality in patients less than 16 years old, between 1985 and 2005. A high failure rate of about 65% was observed in 5- to 12-year-old children with HED-induced teeth agenesis. Implant loss has been attributed to the small jaw size and preoperative conditions [67], however, the medullary bone hyperdensity observed in HED patients should also be encountered as an additional risk factor [68].

4.3 Adolescence - The Early Permanent Dentition Years

During adolescence normal growth changes may influence the skeletal relationships and function that were restored by previous treatment; on the other hand, the opportunity to take advantage of the growth spurt to correct maxillary and mandibular growth defects in HED patients must not be missed [19]. The armamentarium for providing growth modification, function and esthetics at this developmental stage includes RPDs, CDs, fixed partial dentures, functional orthopedic appliances and combinations of them (Fig. 3). As in all preceding treatment phases, composite resin restorations and build-up of existing malformed teeth may also be performed to improve the appearance and provide proper contacts with opposing teeth.

Pre-pubertal maxillary growth in width can be controlled by separating the mid-palatal suture with a removable expansion device [49,69,70]. Therefore, removable appliances, either orthodontic or prosthodontic, equipped with 3-direction expansion screws in the maxilla and 2-direction expansion screws in the mandible are again recommended. Moreover, orthodontic removable appliances may be equipped with active springs for space management (Fig. 4). During the peak of growth rate the removable devices should be activated at a rate of approximately 0.5 mm / week.

The small number of natural teeth as well as the deficient morphology of the edentulous ridges, limits the stability of the denture [33] making it less efficient in opening of the mid-palatal suture. It is therefore essential to maintain good support and retention of the prostheses, either maxillary or mandibular. For this purpose, the prosthodontic means described earlier, i.e. proper fit and controlled extension of the base, functional shaping, lining with soft liners or denture adhesives, are utilized.

The anteroposterior position of the maxilla often must be modified. The anteroposterior position of the maxilla and the mechanisms to influence its growth disturbances are rarely referred in the literature. When referring to anteroposterior position of maxilla is generally to accommodate it in relation to the position of the mandible. Based on their clinical experience, the authors advise that HED patients with marked maxillary deficiency who possess sufficient number of posterior teeth to serve as attachments may benefit from the adjustment of face mask (Fig. 5). Thus, an orthognathic surgery in the end of growth and the skeletal maturity to normalize skeletal jaw relationships may be avoided.

Fig. 3. Intraoral photographs of a 11-year-old boy with HED. (a, b, c) View of the occlusion with excessive overbite and Class III skeletal relationships, (d) the functional orthopedic appliance for the control of the overbite and the improvement of the skeletal discrepancy along with artificial mandibular incisors for the esthetic improvement, and (e, f, g) view of the occlusion with the functional appliance. (Patient of the Postgraduate Clinic of Orthodontics, treated by the authors)

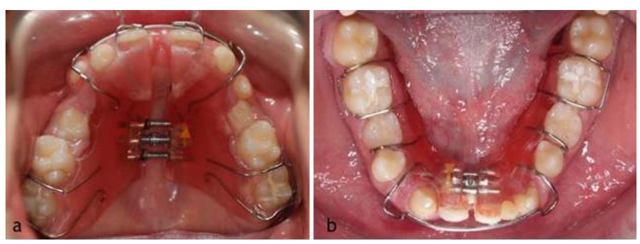

Fig. 4. (a) The maxillary transversal expansion with a removable device equipped with a 2-directional jackscrew, (b) the expansion device at the mandible with artificial mandibular incisors for space management and esthetics. (Patient of the Postgraduate Clinic of Orthodontics, treated by the authors)

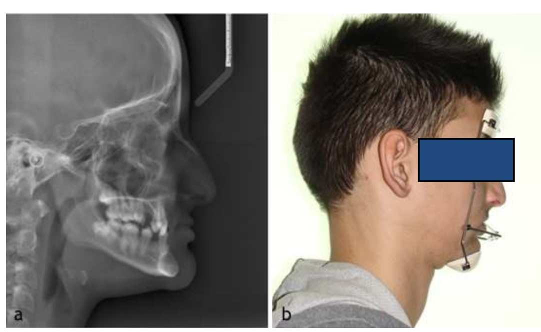

Fig. 5. (a) Lateral cephalometric radiography of HED patient showing the maxillary deficiency. (b) The adjustment of the face mask for the forward displacement of the maxilla. (Patient of the Postgraduate Clinic of Orthodontics, treated by the authors)

Mandibular growth and position can also be modified in the period of puberty. As mentioned before, it is recommended that the removable appliances, either partial or complete, may be constructed with normal overjet but accentuated overbite, in order to prevent the anterior shifting of the mandible. In cases with mandibular growth pattern skeletal Class III, the authors advocate the use of orthopedic appliances, such as chin cup devices to alter the spatial position of the mandible (Fig. 6). In cases with deep bite or over-erupted incisors, functional appliances such as activators, which permit the extrusion of posterior teeth and encourage the development of the dentoalveolar processes, are also indicated [8,9,71].

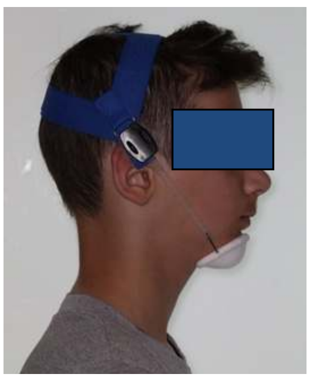

Fig. 6. The chin cup device for the control of the mandibular shift. (Patient of the Postgraduate Clinic of Orthodontics, treated by the authors)

At this stage, as adolescence comes to its end, the permanent treatment plan is gradually built up by the treatment team. In this context, orthodontic treatment is sometimes undertaken to move the remaining teeth towards their final positions (Fig. 7). These are decided according the esthetic needs, and also in order to permanently delineate the spaces for the artificial teeth, either on a tooth-borne or implant-borne prosthesis [8,17]. The patients usually need space closure or opening, distal or mesial movement of existing teeth, and teeth alignment. They may also need overbite reduction and incisors uprighting. Orthodontic fixed appliances allowing the control of tooth movement, improve the distribution of spaces in the dental arch and contribute to the preparation of prosthesis. Removable appliances with jackscrews, functional orthopedic appliances and bite planes all act as stimuli to enhance maxillary and mandibular growth in 3 directions. Along with orthodontic fixed appliances for tooth space alignment and space distribution they guide the modifications necessary for the final rehabilitation in adulthood [70].

4.4 Adulthood

During adulthood growth continues, but in low rates [19]. Small changes affecting the three planes of space are sometimes noticeable and have to be monitored. At this time of residual growth, RPDs or CDs are still constructed and accommodate the changes of the jaws. Orthodontic fixed appliances may also be used in order to stabilize the spaces between teeth, and prevent their overeruption or tilting. The multidisciplinary team discusses the definite treatment options, the patient is presented with the definite restoration alternatives, and the final prosthodontic treatment plan is elaborated, including the necessary pre-prosthetic surgical and/or orthodontic procedures [8,56,70]. For example, orthognathic surgery may be advised, combined with orthodontic intervention, to correct the anteroposterior skeletal discrepancies and to improve the concave profile of HED patients, providing a favorable background for the final prosthodontic rehabilitation [72,73]

Fig. 7. (a, b) Fixed orthodontic appliances for teeth uprighting, alignment and space redistribution in maxilla and mandible. (c) Removable orthodontic appliance in combination with fixed orthodontic appliances to support anterior mandibular artificial teeth. (Patient of the Postgraduate Clinic of Orthodontics, treated by the authors)

Implant supported prostheses are the definite treatment of choice. After growth has been stabilized, implants may be inserted to stabilize the prosthesis, preferably a fixed one, solely or in combination with the natural teeth [4,51,55,56,74-76]. However, it is not in all cases of HED children that implant restorations can be constructed. As mentioned, teeth agenesis is accompanied by severely underdeveloped alveolar bone, which often cannot enfold the necessary number of adequately sized implants. Pre-prosthetic surgery for bone augmentation is most often a necessity, and complicates and prolongs the treatment. Another concern about implant placement is that, because they are inserted in a young age, they are expected to serve for several decades, and there is currently not enough evidence about such long-term function [31,67,77,78].

5. CONCLUSIONS

The rehabilitation of dento-facial malformations in HED patients begins as early as feasible and peaks up by the insertion of a permanent prosthesis at the end of growth. The management through the growing years involves monitoring and, when needed, the modification of growth abnormalities in the 3 levels of space. Close follow-up and team coordination for treatment planning and timing of interventions according to each individual's growth periods are decisive for a successful treatment outcome for HED patients.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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